Ref #	Hits	Search Query	DBs	Default Operator	Plurals	Time Stamp
S4	742	((N-ACETYL-ASPARTATE AMIDOHYDROLASE) OR (N-ACETYLASPARTATE AMIDOHYDROLASE) OR (ASPARTOACYLASE))	US-PGPUB; USPAT; EPO; JPO; DERWENT	OR	ON	2005/09/21 13:32
S5	421	S4 and human	US-PGPUB; USPAT; EPO; JPO; DERWENT	OR	ON	2005/09/21 13:33
S6	361	S5 and ((purification) or (purified) or (isolate) or (isolated))	US-PGPUB; USPAT; EPO; JPO; DERWENT	OR	ON	2005/09/21 13:33
S7	76	((N-ACETYL-ASPARTATE AMIDOHYDROLASE).clm. OR (N-ACETYLASPARTATE AMIDOHYDROLASE).clm. OR (ASPARTOACYLASE).clm.)	US-PGPUB; USPAT; EPO; JPO; DERWENT	OR	ON	2005/09/21 13:32
S8	29	S7 and human	US-PGPUB; USPAT; EPO; JPO; DERWENT	OR	ON	2005/09/21 13:33
S9	25	S8 and ((purification) or (purified) or (isolate) or (isolated))	US-PGPUB; USPAT; EPO; JPO; DERWENT	OR	ON	2005/09/21 13:33

Dialog results pertinent

5/9/8 (Item 1 from file: 35)
DIALOG(R)File 35:Dissertation Abs Online

01283956 ORDER NO: AAD93-10089
CANAVAN DISEASE: ISOLATION AND CHARACTERIZATION OF ASPARTOACYLASE **(MYELINIZATION)**
Author: CASANOVA, JOSE MARIA
Degree: PH.D.
Year: **1992**
Corporate Source/Institution: UNIVERSITY OF ILLINOIS AT CHICAGO, HEALTH
SCIENCES CENTER (0806)
Source: VOLUME 53/12-B OF DISSERTATION ABSTRACTS INTERNATIONAL. PAGE 6236. 120 PAGES
Descriptors: HEALTH SCIENCES, PATHOLOGY; CHEMISTRY, BIOCHEMISTRY; BIOLOGY, NEUROSCIENCE
Descriptor Codes: 0571; 0487; 0317

The aim of the present work was to isolate and characterized brain **aspartoacylase** . By using various fractionation and chromatography procedures bovine brain **aspartoacylase** was **purified** approximately 70,000 fold, to apparent homogeneity. Gel filtration chromatography and SDS-PAGE showed that **aspartoacylase** is a monomeric protein with a molecular weight close to 58-KDa. The partially **purified** enzyme required
divalent cations for activity. The optimal pH was found to be close to 8.0.
The addition of chelating agents, such us EGTA and EDTA, had a severe inhibitory effect on the activity of the crude enzyme preparation. The later two effects were not seen with the **purified** enzyme.
Aspartoacylase
was also shown to be a very stable enzyme, especially in the presence of
low concentration of non-ionic detergents.

Direct biochemical measures of **aspartoacylase** at different levels of
the gray and white matter showed that the enzyme is mainly confined to the
subcortical white matter of the brain. Polyclonal antibodies with a high
degree of specificity against **aspartoacylase** , were shown to react with
the enzyme in the subcortical white matter, following the myelinated tracks.

The importance of **aspartoacylase** has been recently underscored by
the finding that deficiency of this enzyme leads to severe myelin disorders, such as Canavan disease. Therefore it seems that **aspartoacylase**
and N-acetylaspartic acid have important roles in the myelinization of the
CNS.

4/9/2 (Item 2 from file: 155)
DIALOG(R)File 155:MEDLINE(R)

10320501 PMID: 8412017

Canavan **disease: biochemical and molecular studies.**

Matalon R ; Kaul R; Michals K

Research Institute Miami Children's Hospital, FL 33155.

Journal of inherited metabolic disease (NETHERLANDS) **1993** , 16 (4) p744-52, ISSN 0141-8955 Journal Code: 7910918

Publishing Model Print

Document type: Journal Article; Review; Review, Tutorial

Languages: ENGLISH

Main Citation Owner: NLM

Record type: MEDLINE; Completed

Subfile: INDEX MEDICUS

Deficiency of the enzyme **aspartoacylase** and the accumulation of N-acetylaspartic acid lead to a severe leukodystrophy and spongy degeneration of the brain, **Canavan** disease (McKusick 271900). Since our discovery in 1988 of the defect in **Canavan** disease, 144 patients with **Canavan** disease have been diagnosed in our laboratory. Most of these children are of Ashkenazi Jewish extraction. The level of enzyme activity can be used for carrier testing. Prenatal diagnosis has been difficult using the enzyme assay owing to the low activity of **aspartoacylase** in cultured chorionic villus samples or amniocytes. The determination of N-acetylaspartic acid in the amniotic fluid is another parameter for diagnosis; however, the levels may not always be elevated. Bovine and human **aspartoacylase** have been purified in our laboratory. Bovine and human cDNA and genomic clones have been isolated and six exons have been localized. This information is being used for the study of **Canavan** disease at the molecular level. (39 Refs.)

Tags: Female; Pregnancy

Descriptors: ***Canava** n Disease; Amidohydrolases--chemistry--CH; Amidohydrolases--deficiency--DF; Amidohydrolases--genetics--GE; Animals; **Canavan** Disease--diagnosis--DI; DNA, Complementary --isolation and purification--IP; Heterozygote Detection; Humans; Pregnancy; Prenatal Diagnosis

CAS Registry No.: 0 (DNA, Complementary)

Enzyme No.: EC 3.5. (Amidohydrolases); EC 3.5.1.15 (**aspartoacylase**)

Record Date Created: 19931122
Record Date Completed: 19931122

4/9/4 (Item 4 from file: 155)
DIALOG(R)File 155:MEDLINE(R)

08818882 PMID: 2512436
SSIEM Award. Aspartoacylase **deficiency: the enzyme defect in** Canavan **disease.**
Matalon R ; Kaul R; Casanova J; Michals K; Johnson A; Rapin I; Gashkoff P; Deanching M
Department of Pediatrics, University of Illinois, Chicago 60612.
Journal of inherited metabolic disease (NETHERLANDS) **1989** , 12 Suppl 2 p329-31, ISSN 0141-8955 Journal Code: 7910918
Publishing Model Print
Document type: Journal Article
Languages: ENGLISH
Main Citation Owner: NLM
Record type: MEDLINE; Completed
Subfile: INDEX MEDICUS
Descriptors: *Amidohydrolases--deficiency--DF; *Demyelinating Diseases --genetics--GE; Awards and Prizes; Cells, Cultured; Demyelinating Diseases --enzymology--EN; Fibroblasts--enzymology--EN; Genetics, Medical; Heterozygote Detection; Humans; Reference Values; Skin--enzymology--EN; Societies, Medical; United States
Enzyme No.: EC 3.5. (Amidohydrolases); EC 3.5.1.15 (**aspartoacylase**)
Record Date Created: 19900125
Record Date Completed: 19900125

4/9/7 (Item 2 from file: 5)
DIALOG(R)File 5:Biosis Previews(R)

0008096143 BIOSIS NO.: 199243064734
BIOCHEMICAL AND ENZYME CHARACTERIZATION OF CANAVAN **DISEASE**
BOOK TITLE: BONNE-TAMIR, B. AND A. ADAM (ED.). GENETIC DIVERSITY AMONG JEWS: DISEASES AND MARKERS AT THE DNA LEVEL; GOODMAN'S INTERNATIONAL CONFERENCE, ISRAEL, JUNE 1990. XXVIII+460P. OXFORD UNIVERSITY PRESS: NEW YORK, NEW YORK, USA; OXFORD, ENGLAND, UK. ILLUS. MAPS
AUTHOR: **MATALON R** (Reprint); MICHALS K; KAUL R; JOHNSON A B

AUTHOR ADDRESS: RES INST, MIAMI CHILD HOSP, MIAMI, FLA, USA**USA
p140-149 **1992**
ISBN: 0-19-506817-3
DOCUMENT TYPE: Book; Meeting
RECORD TYPE: Citation
LANGUAGE: ENGLISH
DESCRIPTORS: HUMAN JEW TREATMENT PREVENTION GENETIC DISEASE
DESCRIPTORS:
MAJOR CONCEPTS: Anthropology; Enzymology--Biochemistry and Molecular Biophysics; Genetics; Neurology--Human Medicine, Medical Sciences; Sense Organs--Sensory Reception
BIOSYSTEMATIC NAMES: Hominidae--Primates, Mammalia, Vertebrata, Chordata, Animalia
COMMON TAXONOMIC TERMS: Animals; Chordates; Humans; Mammals; Primates; Vertebrates
CONCEPT CODES:
00520 General biology - Symposia, transactions and proceedings
03508 Genetics - Human
05000 Physical anthropology and ethnobiology
10064 Biochemistry studies - Proteins, peptides and amino acids
10806 Enzymes - Chemical and physical
11304 Chordate body regions - Head
12512 Pathology - Therapy
20006 Sense organs - Pathology
20506 Nervous system - Pathology
21006 Psychiatry - Mental retardation
25503 Development and Embryology - Pathology
BIOSYSTEMATIC CODES:
86215 Hominidae